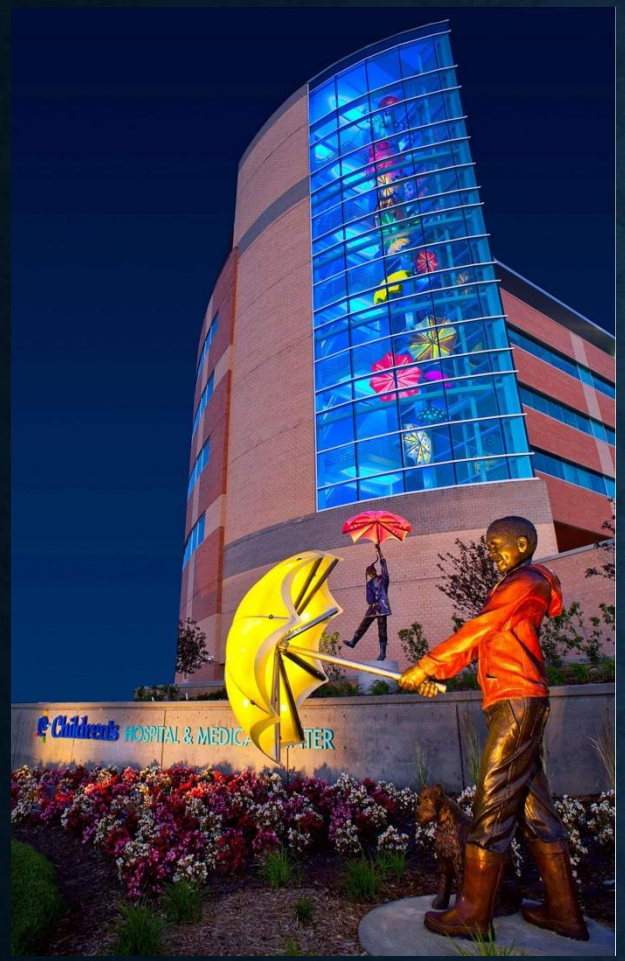

AFTER PEDIATRIC CARDIAC ARREST – WHAT HAPPENS NOW?

Christopher C Erickson, MD, FHRS, FAHA, FACC

Children's Specialty Physicians/UNMC/CUMC

Omaha, NE

UNMC BREAKTHROUGHS FOR LIFE.

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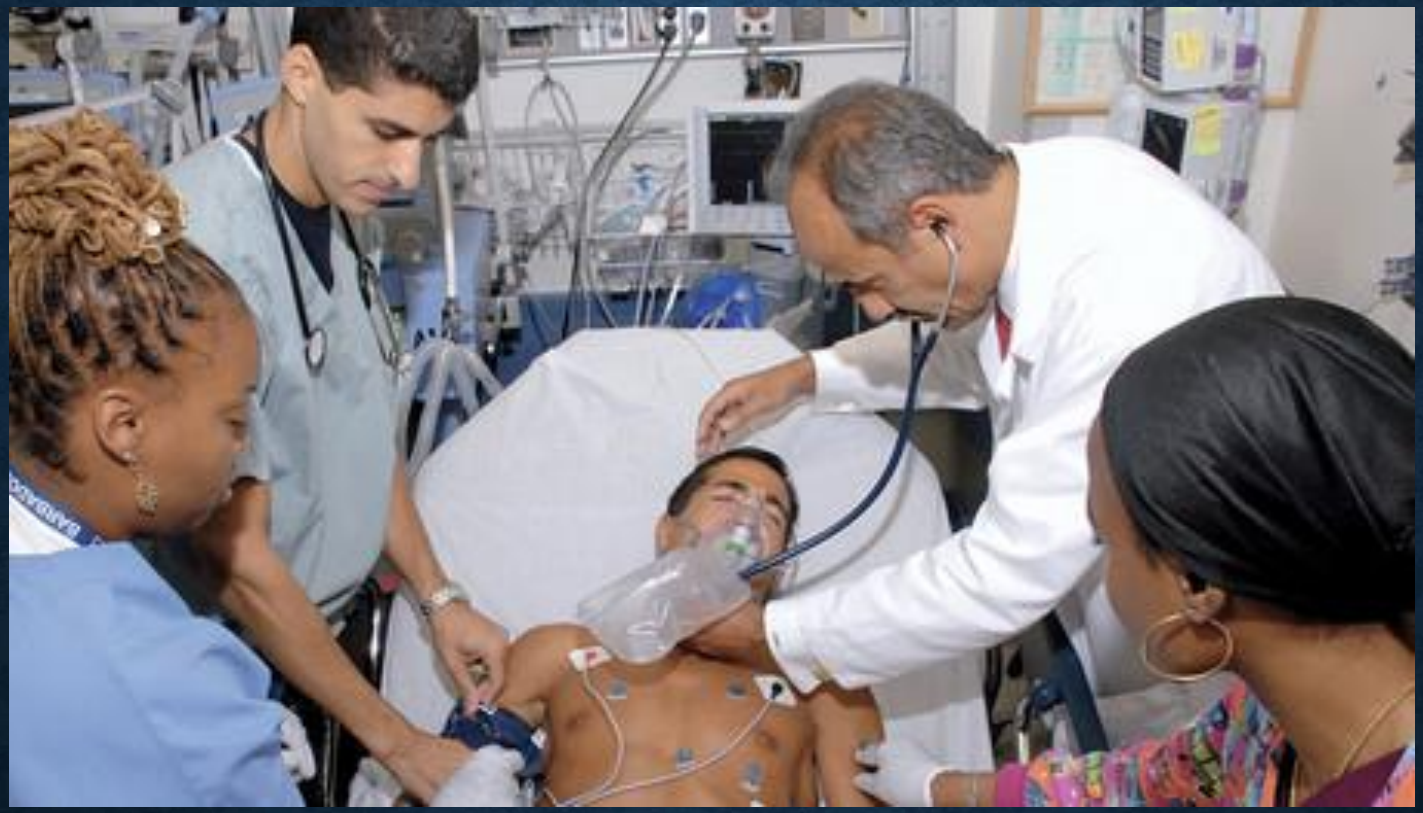
NO DISCLOSURES

OBJECTIVES

- Communicate specific measures need to be done after a pediatric cardiac arrest
  - For those that survive
  - For those that die
- Learn about intermediate to long term post arrest care
- Discuss conditions that predispose pediatric patients to cardiac arrest
- Consider psychiatric issues after cardiac arrest
- Review issues regarding ECG screening

UPON ARRIVAL IN THE ER…



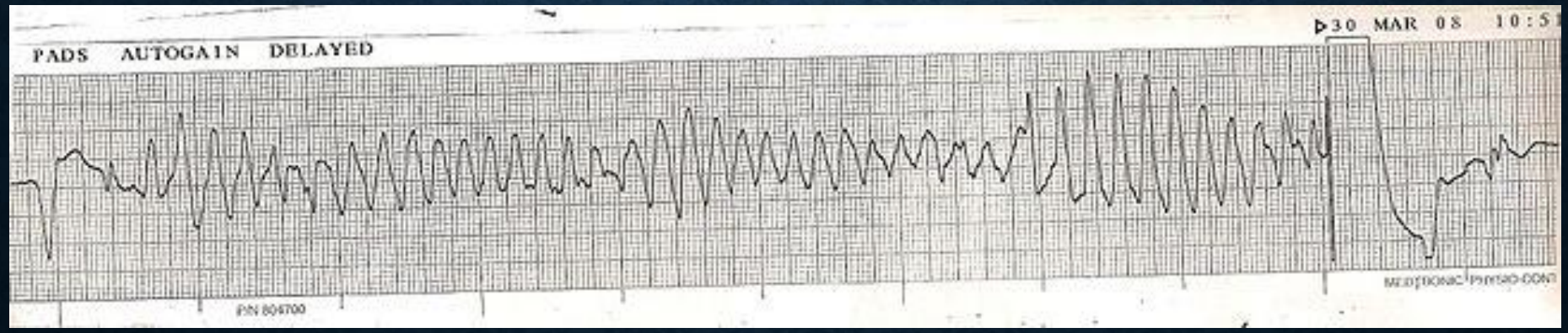
What's Next?

IF THE PATIENT DOESN'T SURVIVE…

- Blood sample should be obtained for molecular autopsy
- History is vital
  - Details of final event by on-the-scene observers is crucial
    - Moment-by-moment detail is necessary
  - History of events preceding final event
    - Syncope, pre-syncope, palpitations
  - Medications
  - Family history
    - Sudden deaths, unexplained accidents
    - Unexplained seizures
    - ICDs or pacemakers in young relatives

IF THE PATIENT DOESN'T SURVIVE…

- Need any AED recordings
  - Documentation of arrhythmias preceding death is very important to potentially aid in diagnosis
- If possible, an echo should be done
- An autopsy is essential to assess for:
  - Cardiomyopathy
  - Myocarditis
  - Coronary abnormalities



AFTER THE FUNERAL…

- Genetic counseling
  - Create family pedigree
    - Creates a “roadmap” of which family members need testing
  - Explain the likely phenotype and genetic implications
  - Explain risk for other family members

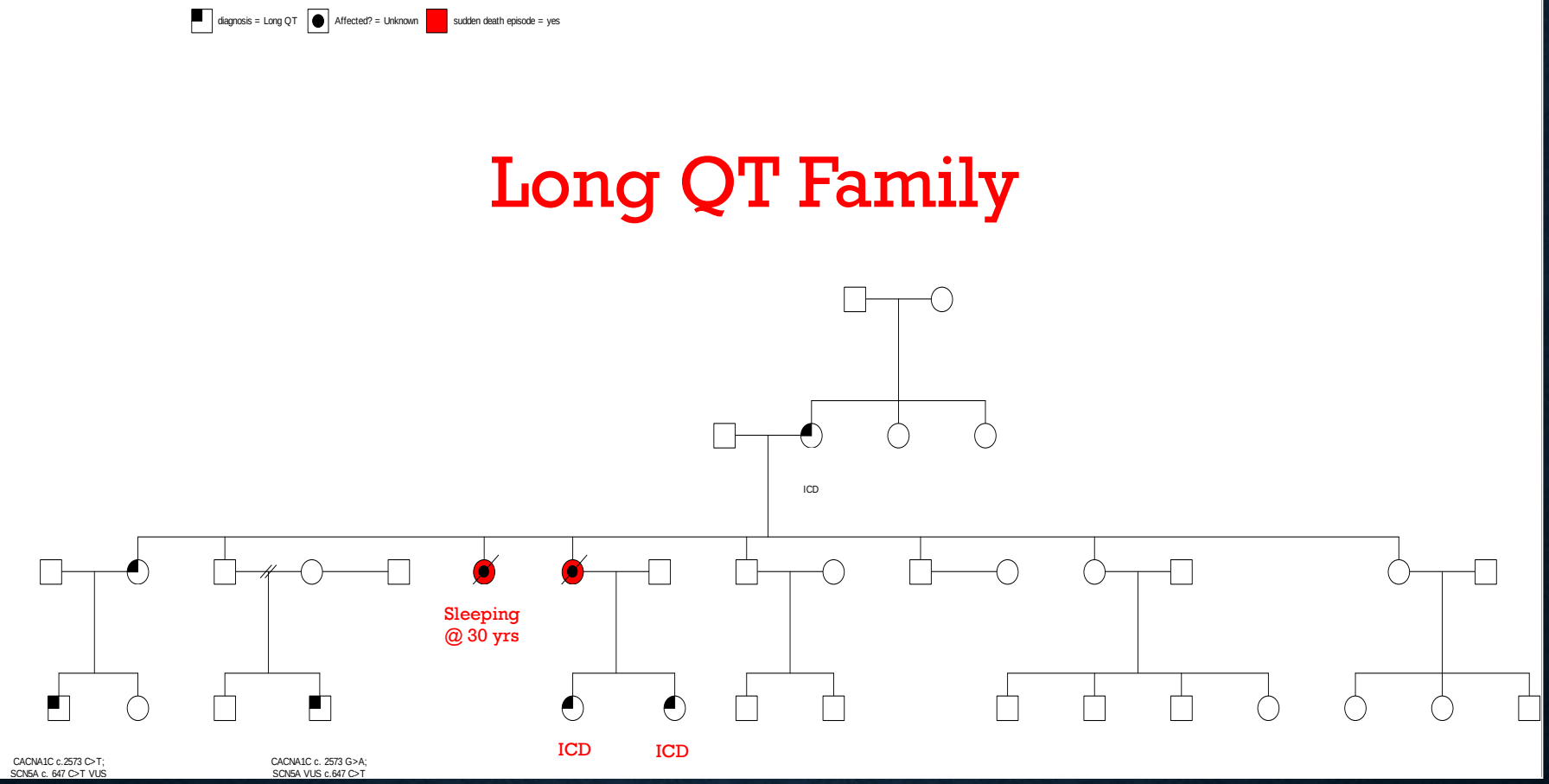
AFTER THE FUNERAL…

- Evaluation of family members
  - Complete history of 1st degree relatives
  - ECGs on mother, father, siblings
  - Possibly will need echo, stress tests, Holters, etc depending on what the phenotype dictates
- Some may need psychiatric counseling

IF THE PATIENT SURVIVES CARDIAC ARREST…

- Full history of events leading to arrest
- Full neurologic evaluation of victim
- If patient is on mechanical ventilation, full assessment may need to be deferred until extubation
- Need to have blood drawn for possible molecular genetic testing
- Detailed history and family Hx is essential
  - Pedigree is created
  - Medications at the time of arrest

Long QT Family



IF THE PATIENT SURVIVES CARDIAC ARREST…

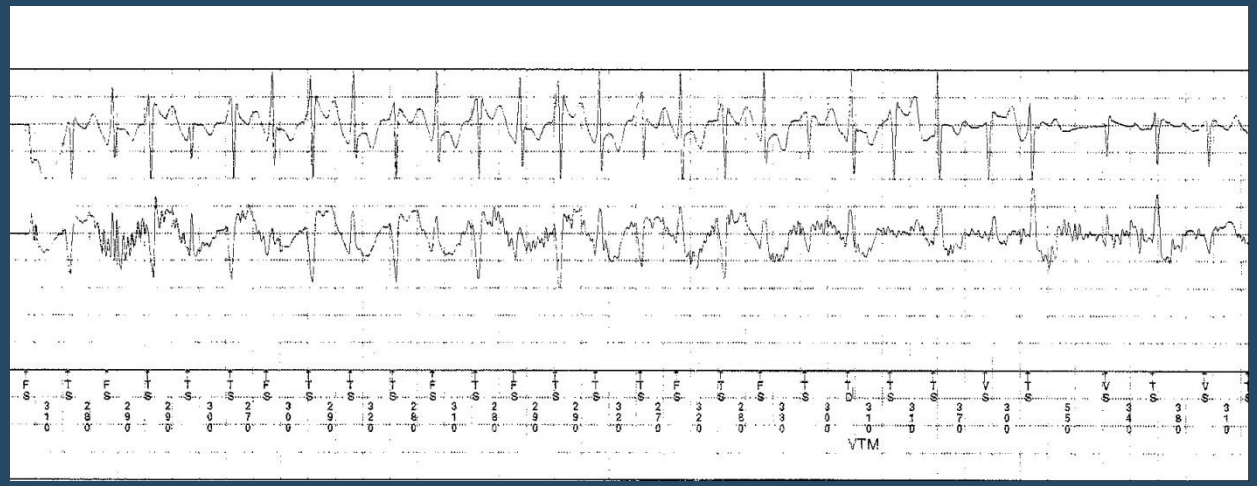
- Psychological evaluation of the victim is necessary
- If no reversible cause is found (drugs, low K+, etc) then the victim is likely to have an ICD implanted (Class I indication)
    - ICD recordings can give some insight as to the cause of the cardiac arrest

CASE

- 13 year old female left in the car with 2 younger brothers while the mother ran into a bank
- The 13 year old got impatient and put the car into drive with the car going over a curb before stopping
- She collapsed and was in cardiac arrest
- Intubated and resuscitated with *near* full neurologic recovery
  - Was a straight A student
  - Now getting C's with some memory impairment

CASE

- Patient has ICD implanted
- Patient and family evaluation including ECGs and epinephrine challenge is negative
- 1 year later she presents for routine ICD follow-up
- This recording is found on her ICD:



Bidirectional Ventricular Tachycardia highly suggestive of Catecholaminergic Polymorphic Ventricular Tachycardia

CASE

- Pt found to have Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT)
- Genetic testing was positive for a mutation on RYR2
- One of two brothers had positive genetic testing for RYR2
  - Subsequent clinical testing was positive for polymorphic VT
  - The brother was started on a beta blocker
  - He decided to stop taking the beta blocker
  - Had a cardiac arrest in a swimming pool
  - Resuscitated and now has an ICD

CONGENITAL HEART DISEASE

- Defects that are present at birth
  - Some corrected
  - Some are only palliated
- Often associated with arrhythmias
  - Cardiac arrest occurs at a higher rate than general population

COMMON DEFECTS

- Holes
  - Atrial Septal Defects
  - Ventricular Septal Defects
  - AV Canal Defects
- Valves
  - Aortic or Pulmonary Stenosis
  - Tricuspid Atresia
  - Mitral Atresia
  - Pulmonary Atresia
  - Aortic Atresia
  - Ebstein's Anomaly

- Great Vessels
- Transposition of Great Arteries (D and L types)
  - Truncus Arteriosus
  - Double Outlet Right Vent
  - Double Inlet Left Vent
  - Coarctation of the Aorta
- Great Veins
  - Total or Partial Anomalous Pulmonary Venous Connections
  - Absent Superior Vena Cava
  - Dual Superior Vena Cavae
  - Interrupted Inferior Vena Cava

PROBLEMS IN PATIENTS WITH REPAIRED CHD

- Residual valve narrowing or leakage
- Decreased ventricular function
- Residual defects
- Single ventricle
  - Surgical palliation - routes blue blood directly to the lungs and only red blood back to the atria/ventricles
    - The patient's only ventricle has to be devoted to pumping blood to high pressure dependent systemic arteries
    - Therefore no ventricle pumps blood to the low pressure lungs

Unrepairable right to left shunts (patients are blue)

- Rhythm disorders

Coronary Artery Anomalies

- Usually Anomalous Left Coronary Artery presents in infancy with angina symptoms and cardiomyopathy
- Presenting symptom of other forms is often sudden death or exertional angina
- Other forms can include:
  - Left main coronary artery arising from the right sinus of Valsalva (passes between the aorta and pulm artery
  - Coronary ostial stenosis
  - Single coronary ostium
  - Intramural origin of the LAD
  - Anomalous Right Coronary Artery (presents in 2nd to 3rd decades)

Coronary Artery Anomalies

"Pistol" Pete Marovich

- Played high school career
- Starred at LSU
  - NCAA scoring leader for 3 years averaging 43.8/44.2/44.5 pts per game
- Played for New Orleans Jazz until sidelined by injury
- Died suddenly during a pick up game of basketball at age 40
  - Found to have a single coronary artery

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Coronary Artery Anomalies

- Diagnosis can be difficult – echo/cath
- Only treatment is surgical – re-implantation or rerouting of the anomalous coronary artery
- May need to treat for dilated cardiomyopathy including anti-CHF and anti-arrhythmic therapies

CARDIOMYOPATHIES

- Hypertrophic Cardiomyopathy (HCM)
- Dilated Cardiomyopathy
  - Multiple causes
- Restrictive Cardiomyopathy
- Arrhythmogenic Right Ventricular Cardiomyopathy
- Left Ventricular Non-compaction

Hypertrophic Cardiomyopathy

Hank Gathers - HCM

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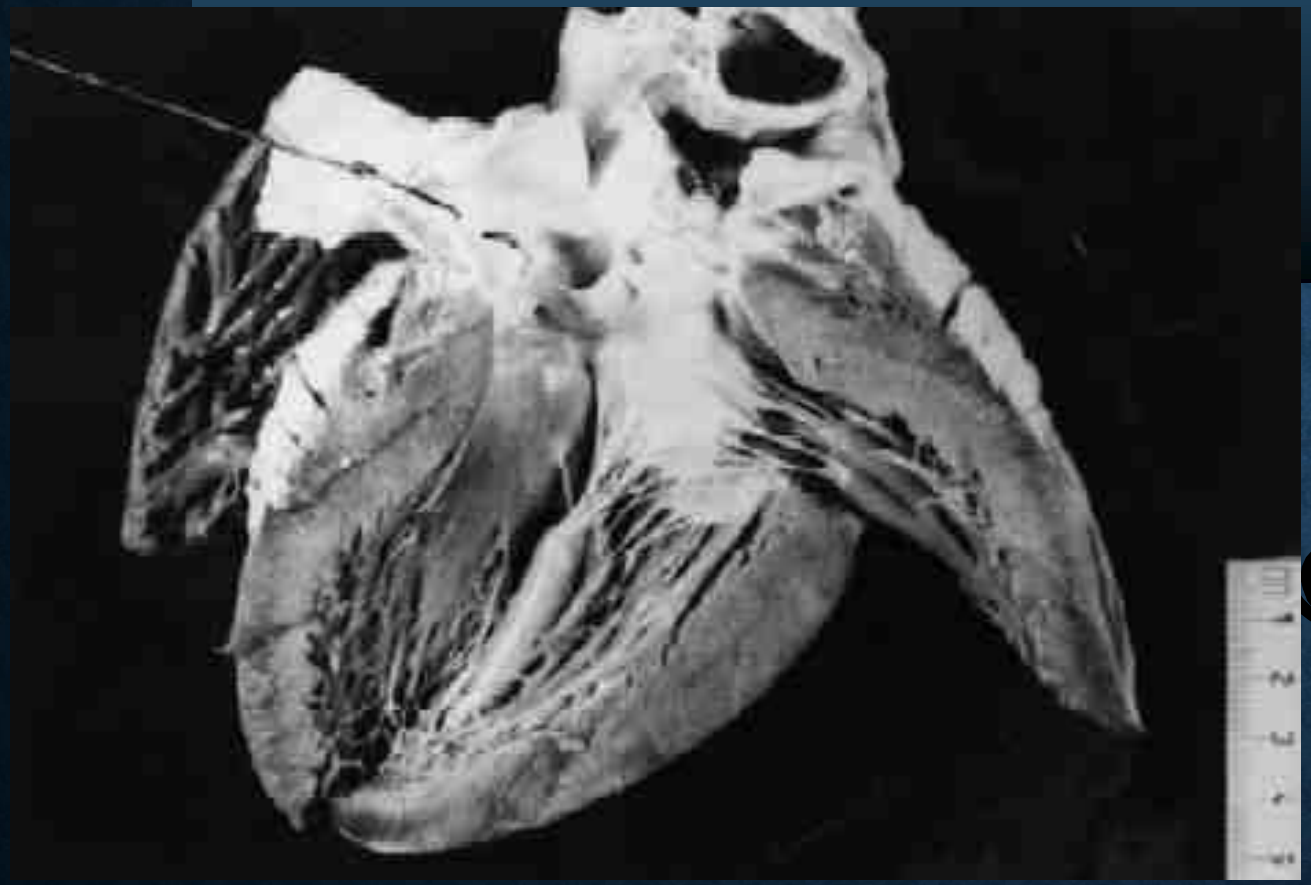
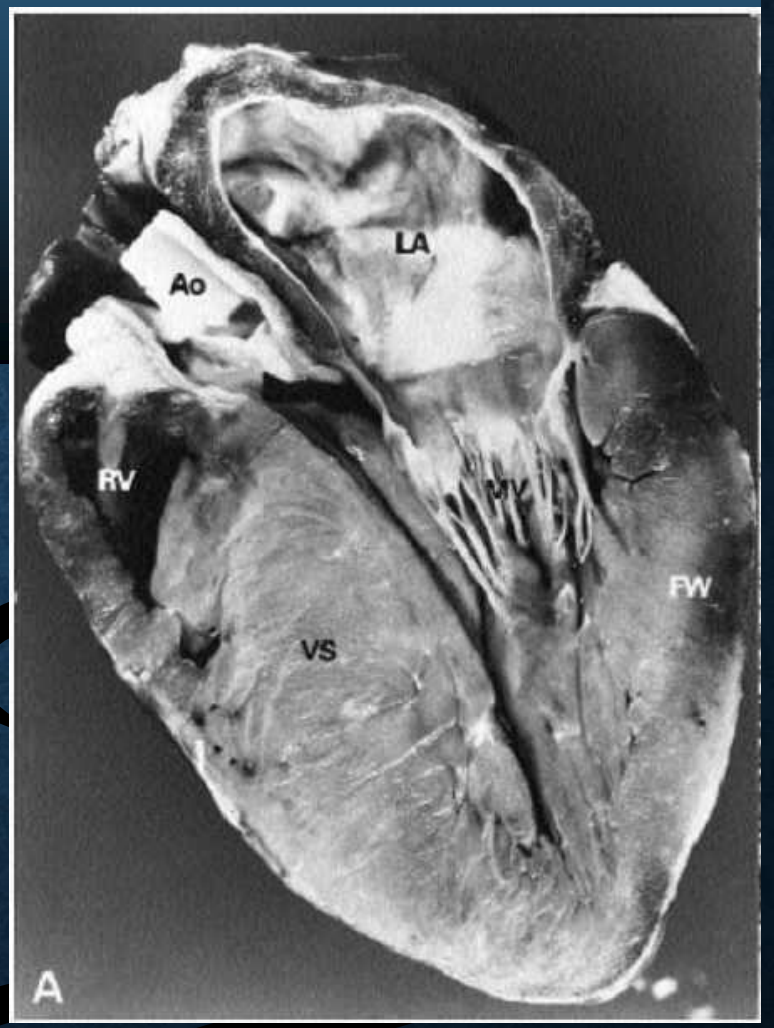
NORMAL VS HCM



Basso



Maron

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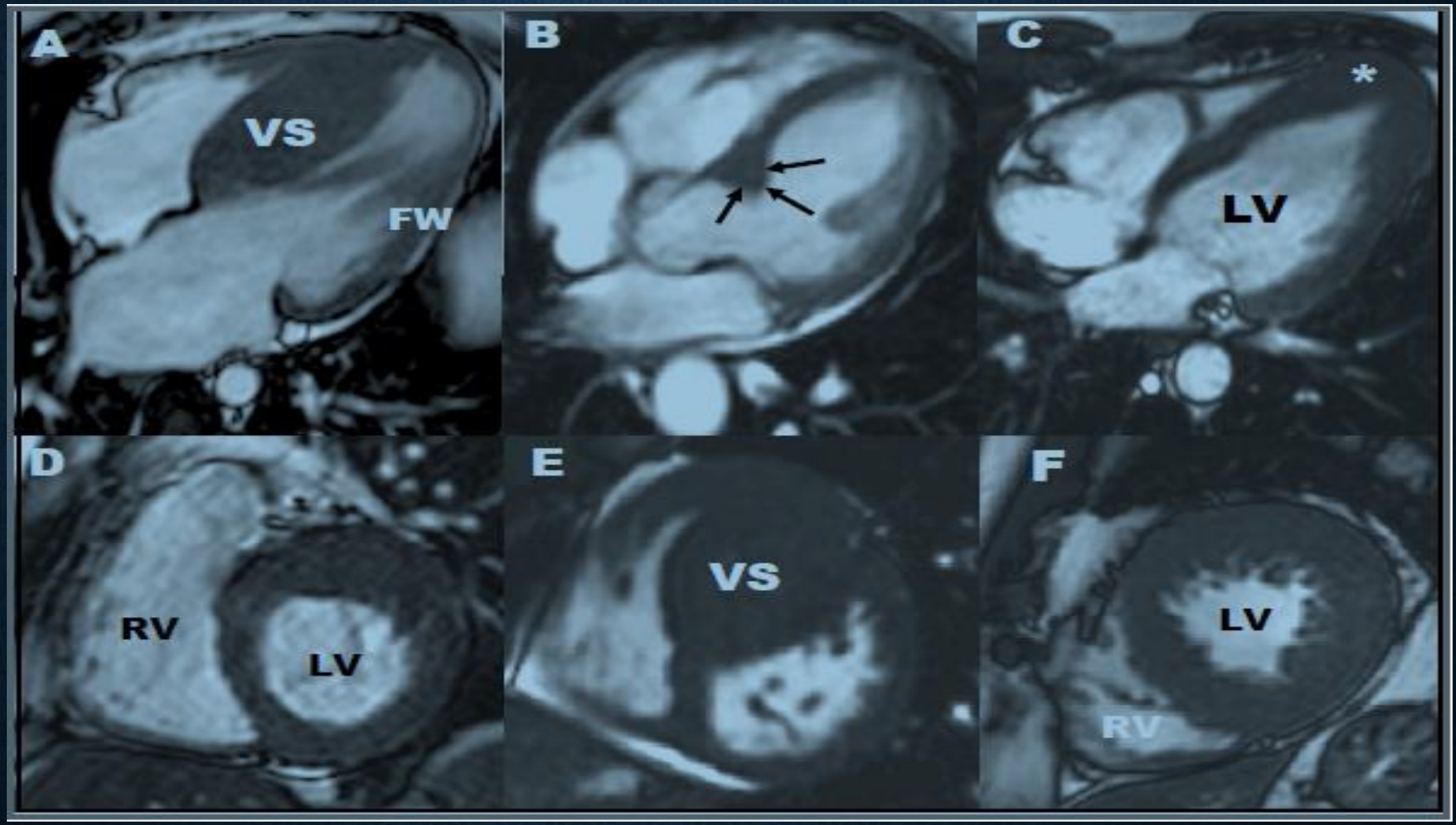
HYPERTROPHIC CARDIOMYOPATHY

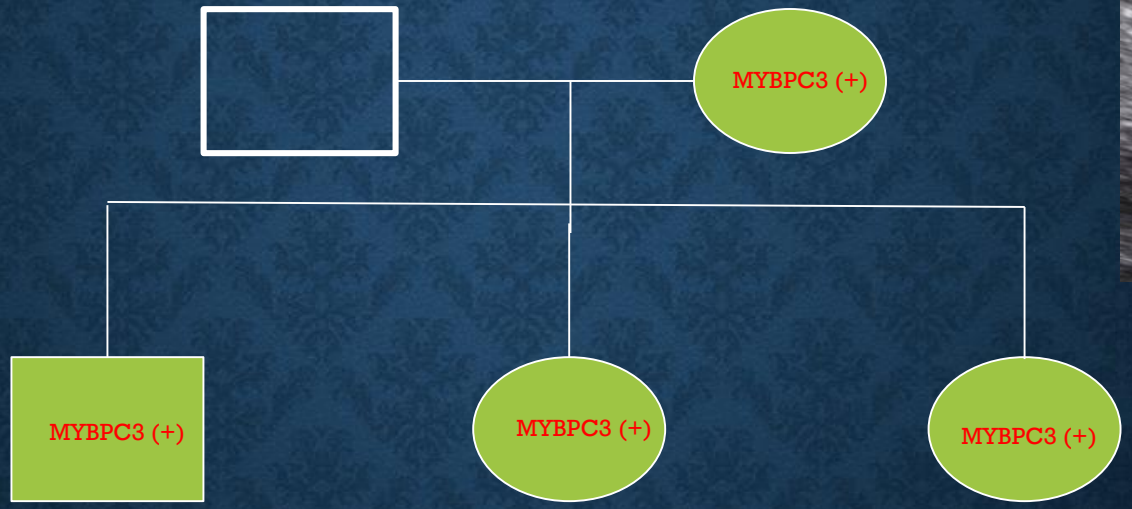
- Abnormal thickening of the heart
- Autosomal dominant inheritance
- Can be due to abnormalities in muscle proteins or storage diseases
- Many genetic expressions
- Some patients have risk factors for sudden death
- HCM can result in heart failure

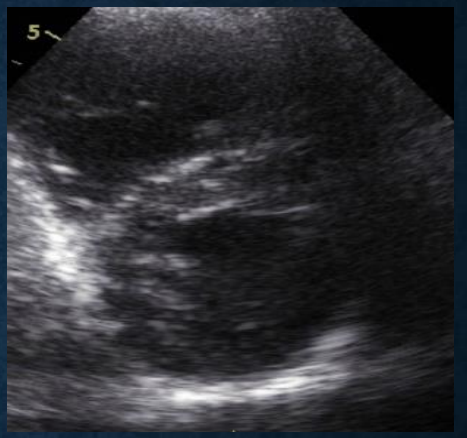
MRI



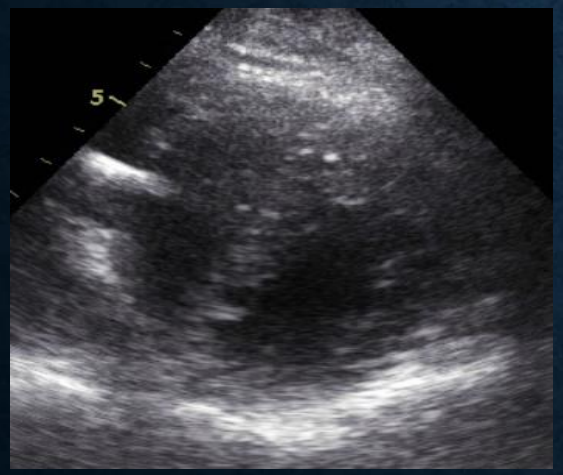
Maron, HCM Summit,2013

PEDIGREE OF HCM (MYBPC3) FAMILY – DIFFERENT EXPRESSION

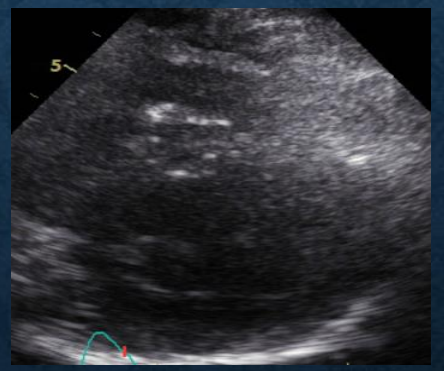




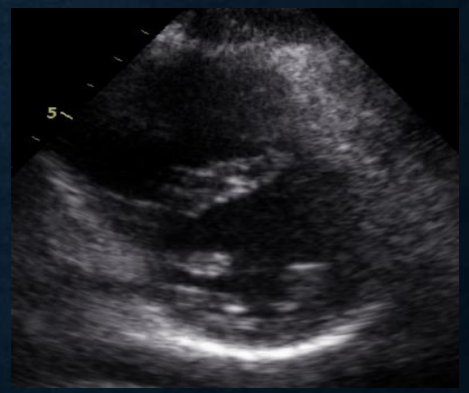
Septum = 19 mm



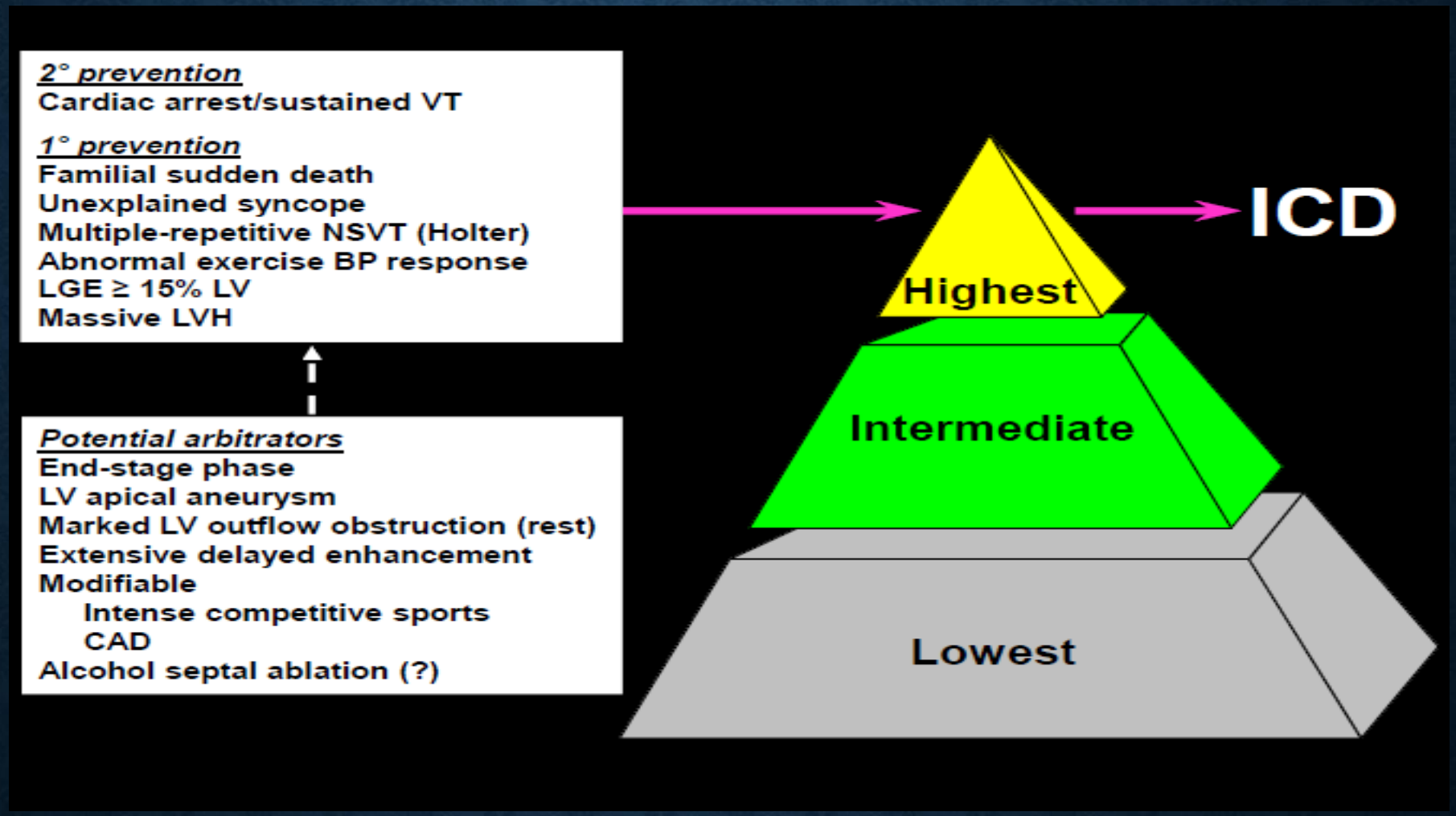
Septum = 34 mm



Septum = 12 mm



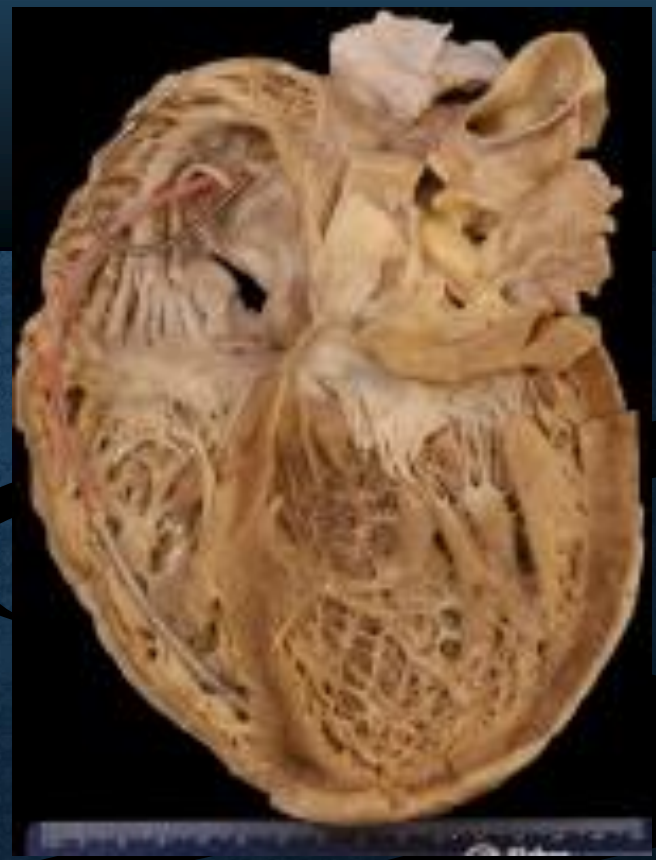
Septum = 8 mm



Maron, HCM Summit 2013

DILATED CARDIOMYOPATHY

- Caused by a variety of etiologies:
  - Familial
  - Viral
  - “Non-ischemic”
  - Storage disease
- Patients can develop:
  - Heart failure
  - Lethal and non-lethal arrhythmias



Burke

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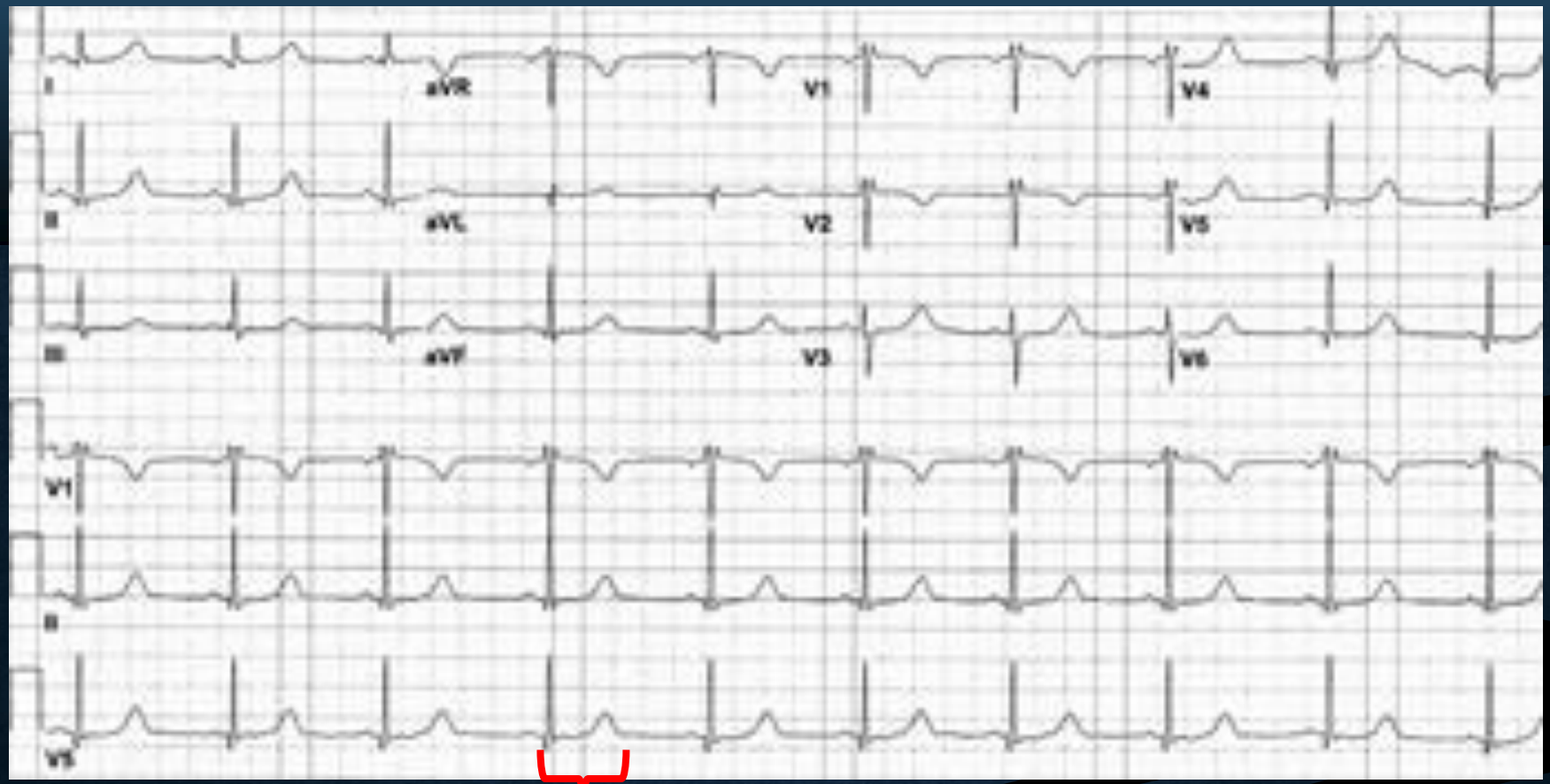
CHANNELOPATHIES

- Cardiac disease at the cellular level causing potential for life threatening arrhythmias
- Involves the opening (channel or pore) into cardiac cells that allows for movement of Na+, K+, and Ca++ in and out of the cells. Abnormal formation of the opening can result in the rhythm problem.
- Typically due to genetic abnormality
  - Therefore can be passed along to the patient's children
- More difficult identify since the heart is structurally normal most of the time therefore they don't have symptoms of poor cardiac output

CHANNELOPATHIES

- Long QT Syndrome – hallmark channelopathy
- Catecholaminergic Polymorphic Ventricular Tachycardia
- Brugada Syndrome
- Short QT Syndrome
- Idiopathic ventricular fibrillation

LONG QT SYNDROME



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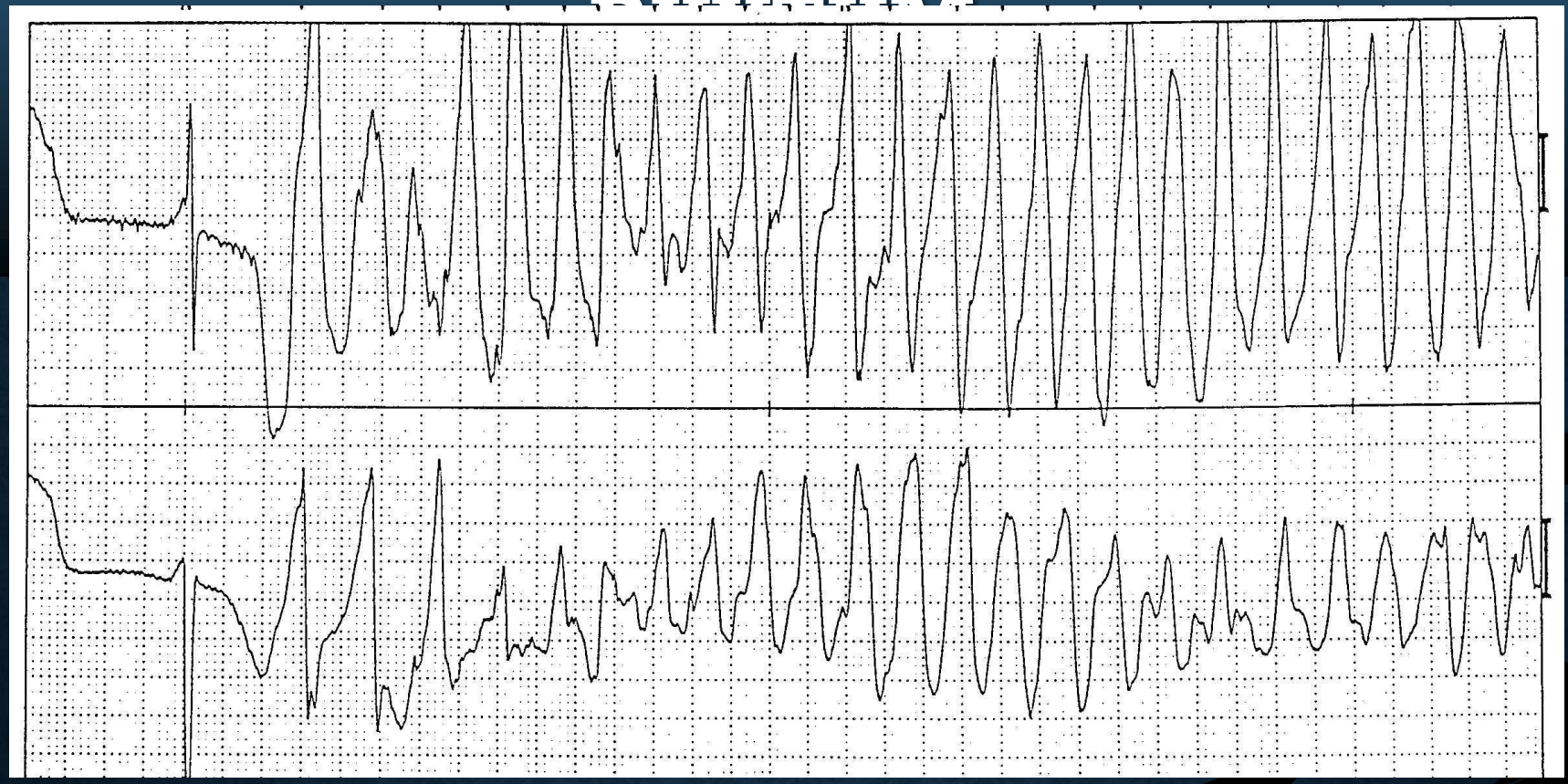
TORSADE DE POINTES – DANGEROUS RHYTHM



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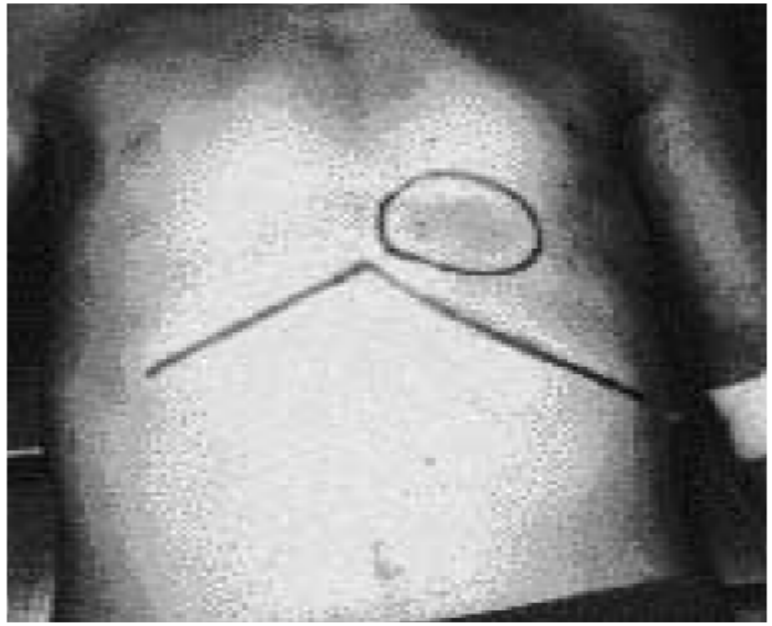
CHANNELOPATHIES AND ATHLETICS

- ACC guidelines restrict these patients to sedentary sports - curling, bowling, and golf
- New data showing the incidence of cardiac events is low in Long QT patients participating in sports
- Many patients will be on beta blockers
  - Side effects could affect athletic performance
- Syncope (fainting) during exercise is a potentially ominous sign
  - Athlete should stop participating immediately
  - The athlete should be reassessed by their electrophysiologist

COMMOTIO CORDIS

- Cardiac arrest caused by impact to the chest
- Earliest description in the literature in 1953
- First series of 4 pathology cases reported in 1984
- Maron reported 25 cardiac arrest victims from 3 – 19 years
  - 7 had protective chest padding

COMMOTIO CORDIS



A

B

C

Figure 2. A 15-Year-Old Victim of Blunt Nonpenetrating Impact to the Chest Delivered by a Hockey Puck (Subject 17).

The incident occurred during a competitive interscholastic hockey game in which the boy rose from a prone position after a melee in front of the goal, raised his arms above his head, and was struck in the chest at close range by a puck from a forehand shot toward the goal. Panel A shows the boy with his protective chest gear removed. A relatively small mid-precordial contusion (3 cm in diameter) produced by the impact of the puck is present just to the left of the sternum, demarcated by the blue circle; the blue line delineates the inferior margins of the rib cage. Panel B shows the plastic-and-foam chest and shoulder protector in its proper position as worn by the victim (with arms at his sides); here the contusion appears to be covered by the chest protector. In Panel C, the arms of the victim are raised (to simulate their position at the moment of the accident, when the victim was attempting to break the flight of the puck), elevating the chest and shoulder padding and leaving the area of impact unprotected.

From Maron, NEJM, 1995

COMMOTIO CORDIS

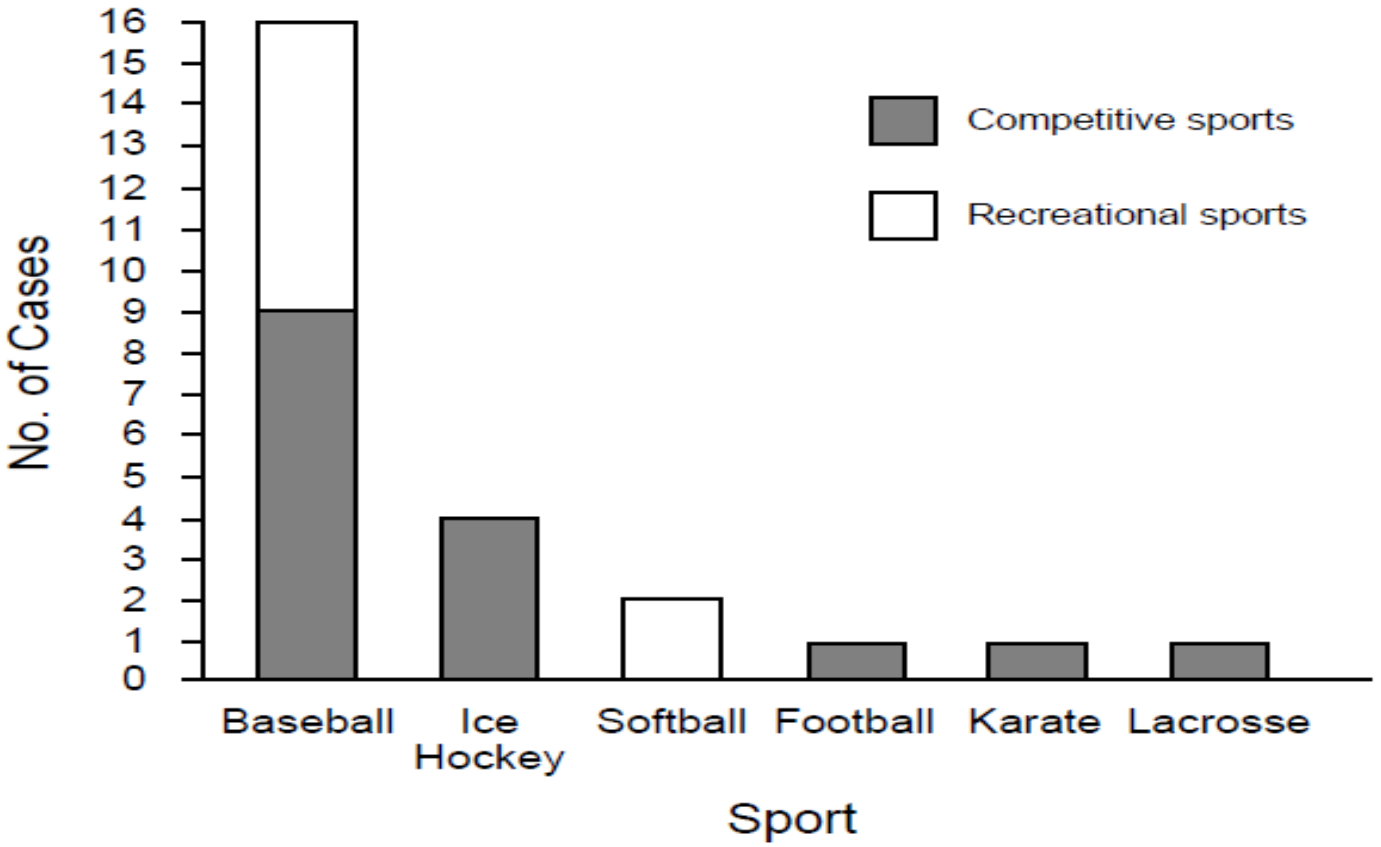


Figure 1. Participation in Competitive and Recreational Sports at the Time of Sudden Cardiac Death Induced by Blunt Impact to the Chest.

From Maron, NEJM, 1995

COMMOTIO CORDIS

From Maron, NEJM, 1995

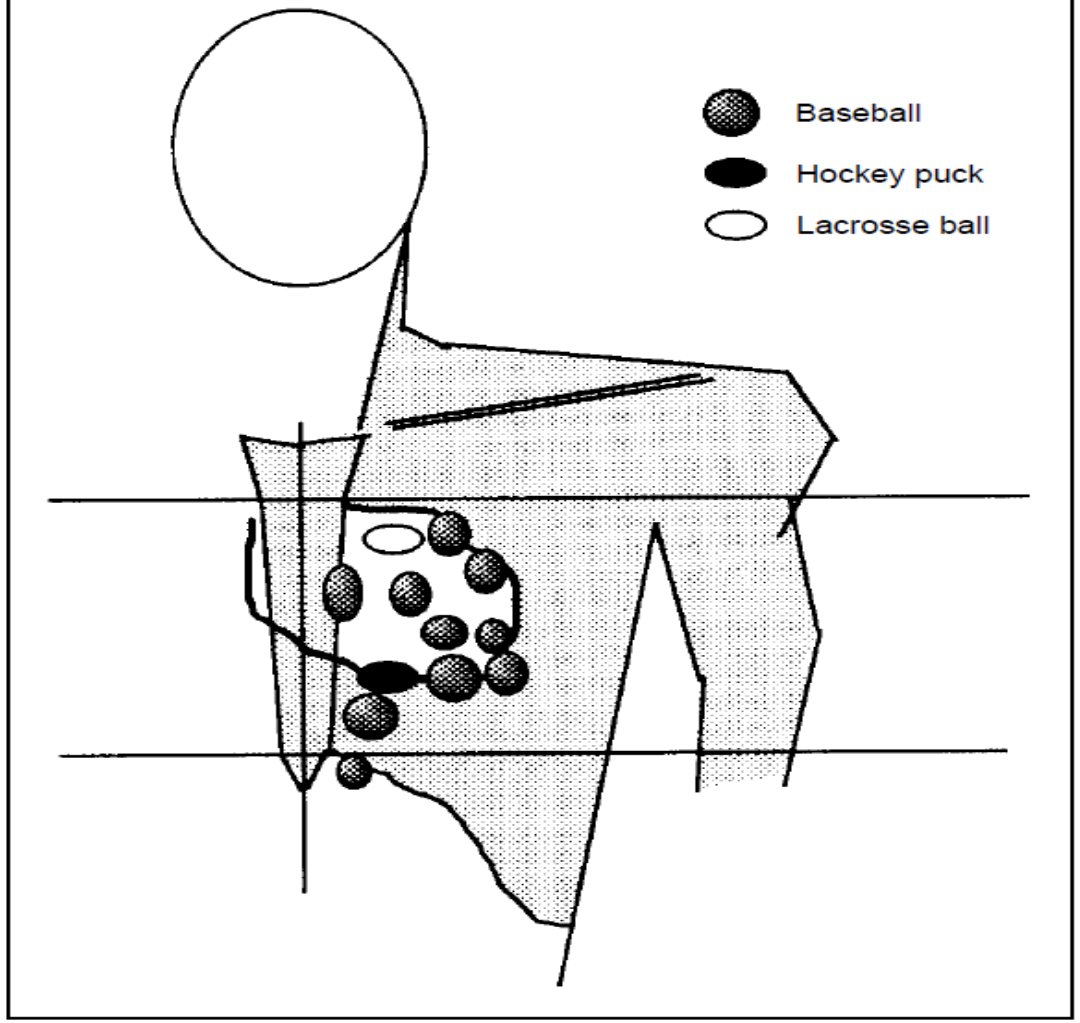


Figure 3. Schematic Representation of the Locations of Impact Points (Contusions) Judged to Have Been Produced by Baseballs (N = 10), a Hockey Puck, and a Lacrosse Ball on the Anterior Chest Walls of 12 Victims of Commotio Cordis.
The estimated contour of the heart is indicated by the heavy line.

COMMOTIO CORDIS

From Link, Circ EP, 2012

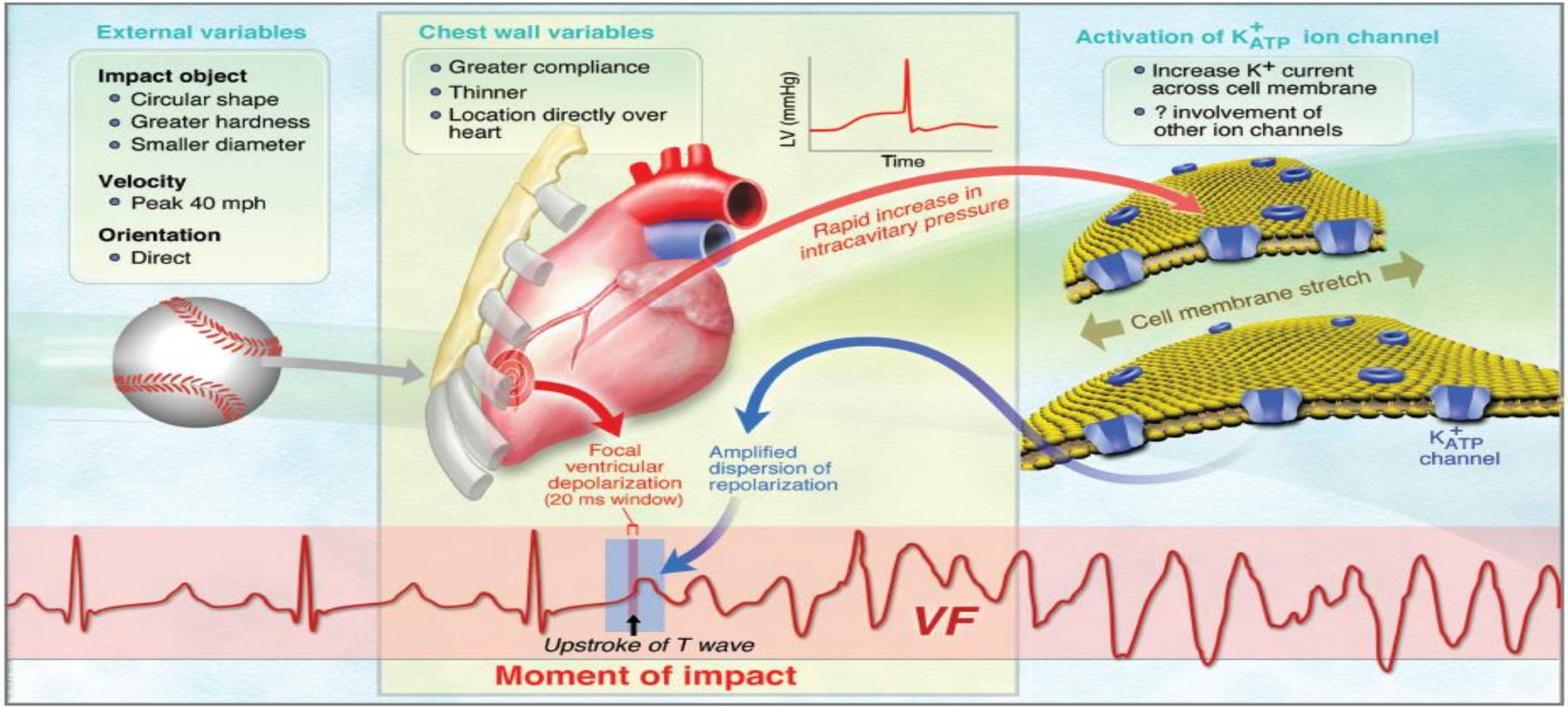
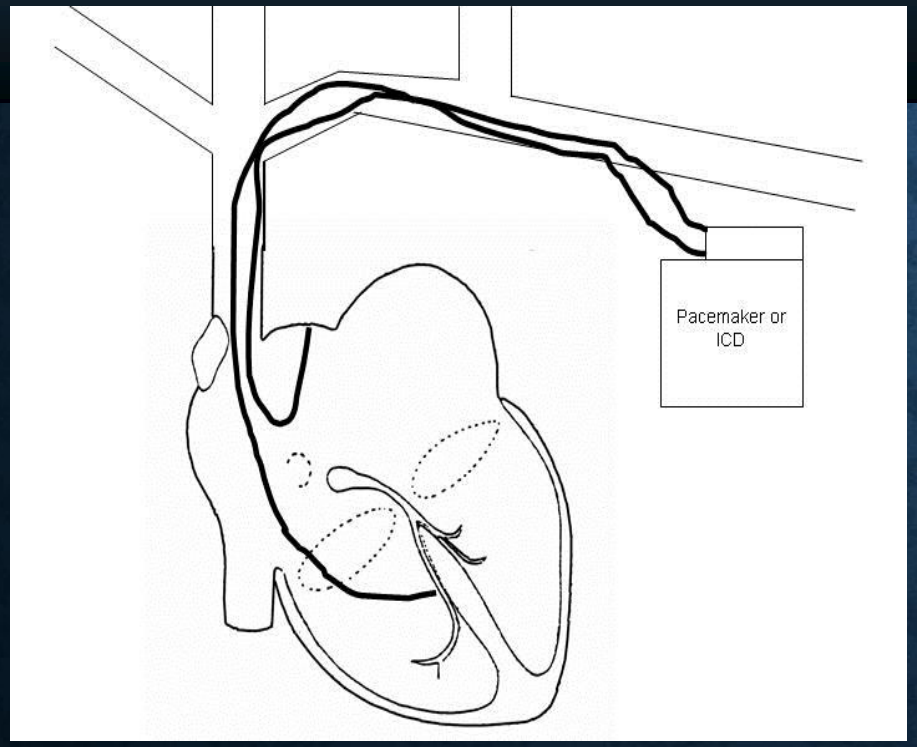


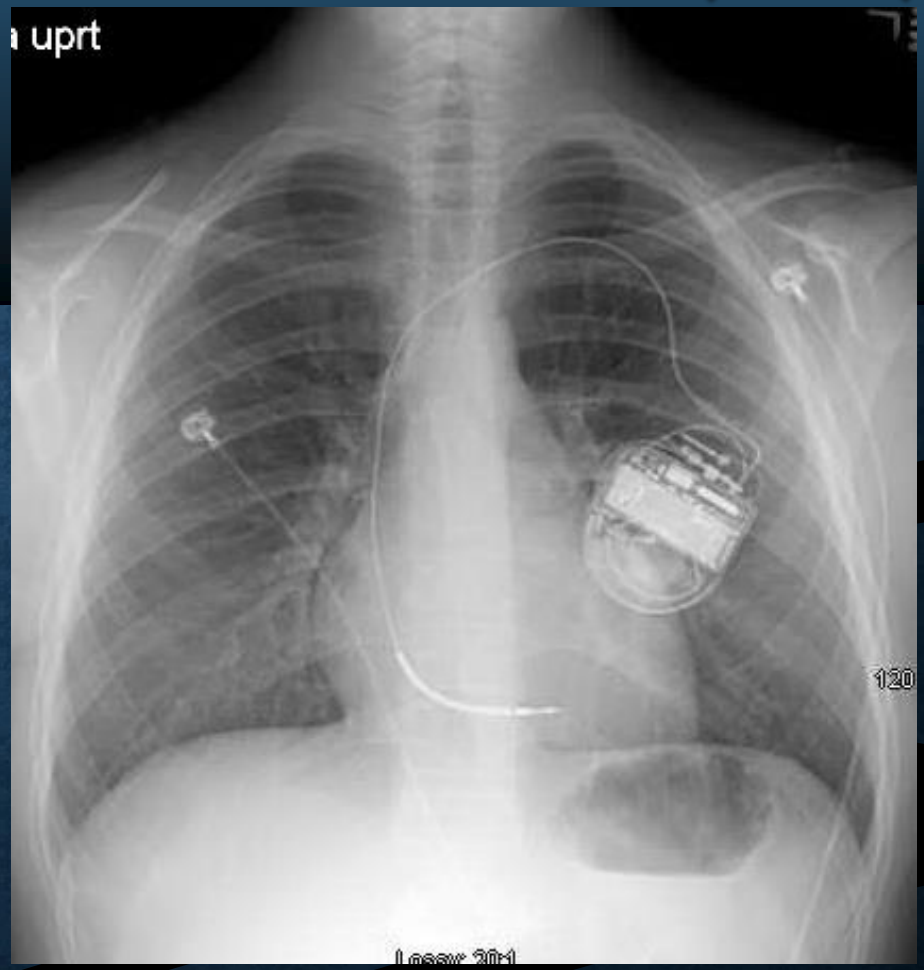
Figure 3. The confluence of variables and a proposed mechanism necessary for commotio cordis to occur. Important impact-object variables are shape, hardness, diameter, and velocity. Human characteristics are the pliability of the chest wall, impact timing, location and orientation of blow, and individual susceptibility, likely carried in ion channels involved in repolarization. LV indicates left ventricle. Reprinted from the *Journal of Cardiovascular Electrophysiology*, with permission.[15]

PATIENTS WITH PACEMAKERS AND ICDS

- Pacemakers & ICDs are usually implanted in the left upper chest with wires entering the vein just under the clavicle
- Generally restricted only from football and martial arts
  - Concern that leads (wires) would fracture where they enter the device
  - More restrictions may be from the disease itself (i.e. channelopathy)

TRANSVENOUS IMPLANTABLE CARDIOVERTER-DEFIBRILLATOR (ICD)





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Controversy of ECG screening in Pediatric Patients

WHAT AN ECG CAN DETECT

- HCM
- Anomalous coronary artery
- Long QT Syndrome
- Brugada Syndrome
- WPW
- Dilated cardiomyopathy
- ARVC
- LVNC

What an ECG can miss

- HCM
- Anomalous coronary artery
- Long QT Syndrome
- Brugada Syndrome
- WPW - Subtle pre-excitation
- Dilated cardiomyopathy
- ARVC
- LVNC
- Exercise induced VT
- CPVT
- Marfan Syndrome

A LOOK AT THE DATA…

- 1997 – Nevada H.S. study (Fuller, Med Sci Sports Exer)
  - 5615 young athletes screened
    - 5033 with Nl Hx/PE/ECG
    - 582 with AbNl Hx/PE/ECG >> echo
  - 99.6% approved for athletics
    - Not approved = 1 severe AI, 5 severe HTN, 16 arrhythmias/conduction abnormalities

A LOOK AT THE DATA…

- 1998 – Italy (Corrado, NEJM, 1998)
  - Athletes required to have annual clinical eval (Hx/PE/ECG/step test) in order to compete since 1971
  - 33,735 athletes (< 35 y.o.) over 17 years
    - 269 SDs
      - 220 non-athletes – (0.75 per 100,000 population/yr)
      - 49 athletes – (1.6 per 100,000 population/yr)
        - Most common C.O.D. = ARVC 22%, atheroscler CAD 18%, anom cor 12%
        - HCM – only 1 death
        - PRE-participation “screen” >> 10 had ST abnormalities, 8 had vent arrhy.
    - 1058 disqualified (58.7% cardiac)
      - 38% rhythm and conduction abnl, 27% HTN, 21% valvular diseases, 3.5% HCM

REDUCTION IN SCD IN ATHLETES AFTER "ECG SCREENING" IMPLEMENTED IN ITALY

JACC Vol. 52, No. 24, 2008
December 9, 2008:1981–9

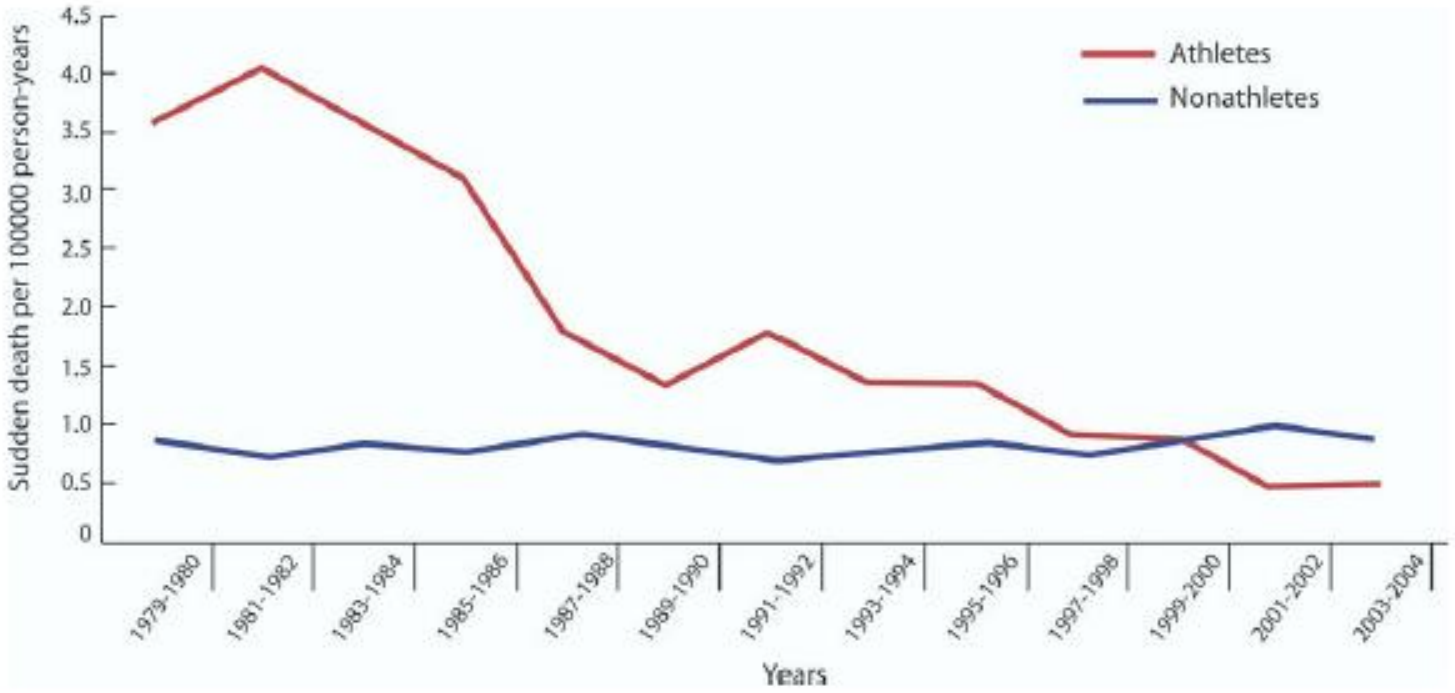

Figure 3 Annual Incidence Rates of Sudden Cardiac Death Among Screened Competitive Athletes and Unscreened Nonathletes in the Veneto Region of Italy From 1979 to 2004

Modified from Corrado et al. (23).

SUDDEN DEATH IN ISRAELI ATHLETES

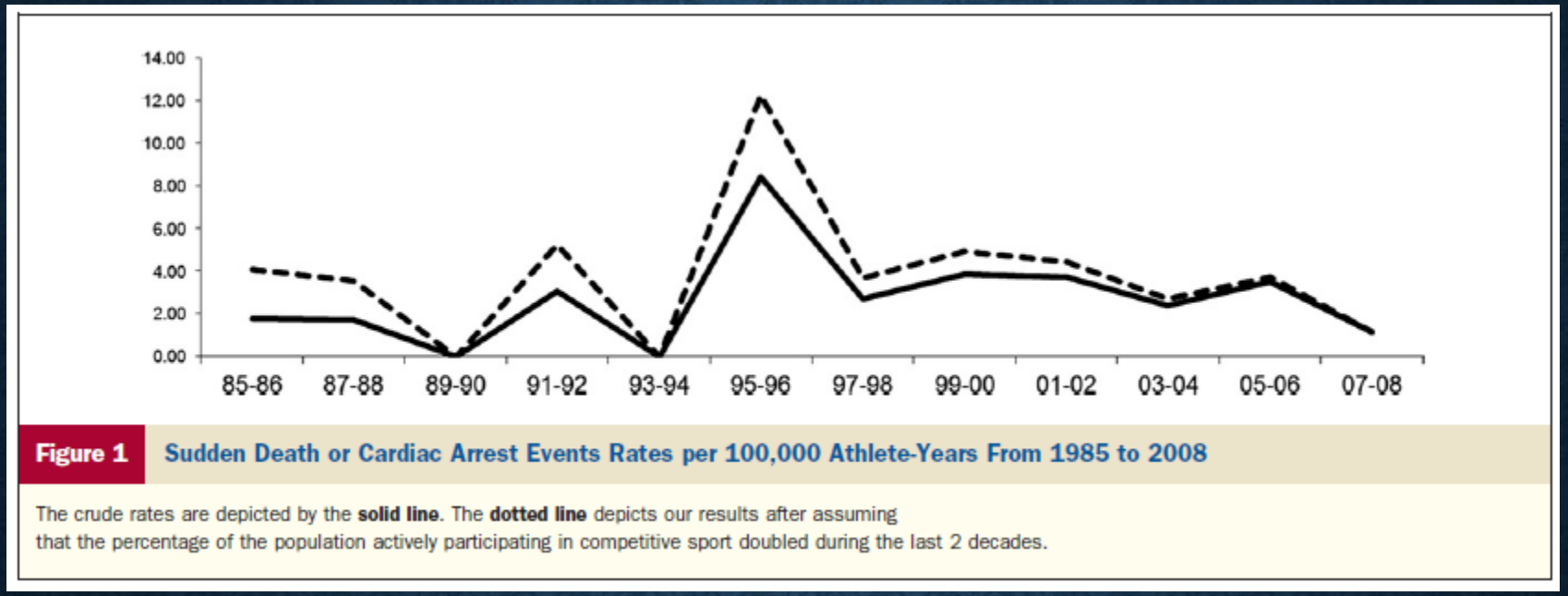

Figure 1 Sudden Death or Cardiac Arrest Events Rates per 100,000 Athlete-Years From 1985 to 2008

The crude rates are depicted by the **solid line**. The **dotted line** depicts our results after assuming that the percentage of the population actively participating in competitive sport doubled during the last 2 decades.

- 24 year study from newspaper accounts
- 1997 – Israel passed law requiring athletes to have annual ECG (all athletes) and exercise testing (every 4 years for those 17-34 and yearly for those 35 and over)
- Incidence of SD was 2.54/100,000 athlete-yrs before ECG law and 2.66 after the law

From Steinvil, JACC, 2011

COMPARISON OF SUDDEN DEATH IN ATHLETE STUDIES

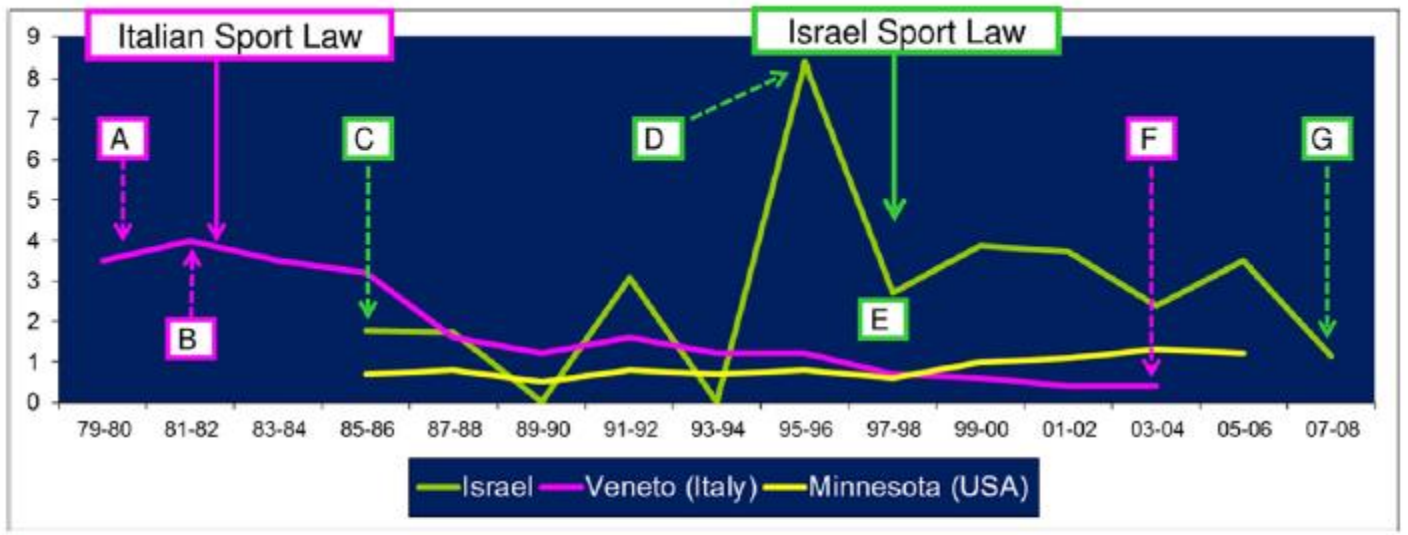

Figure 2 **Annual Incidence of Sudden Cardiac Death Expressed per 100,000 Person-Years in the 3 Studies Evaluating the Effects of Screening on the Mortality of Athletes Over Time**

The Italian study (4) **(pink graph)** concluded that electrocardiography (ECG) screening (started in 1982) significantly reduced the incidence of sudden cardiac death by comparing the sudden death in the 2-year pre-screening period **(A to B)** with the post-screening period **(B to F)**. The present study is depicted by the **green graph**. We compared the 12 years before screening **(C to E)** with the 12 years after the onset of mandatory ECG screening **(E to G)**. Had we limited our comparison of the post-screening period to the 2-year period preceding the enforcement of screening in Israel (**D to E** vs. **E to G**, as performed in the Italian study), we would have concluded erroneously that screening saved lives of athletes in Israel. The study from Minnesota (19) **(yellow graph)** shows a low mortality rate in a population of athletes not undergoing systematic ECG screening.

From Steinvil, JACC, 2011

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SCIENTIFIC STATEMENT

Assessment of the 12-Lead Electrocardiogram as a Screening Test for Detection of Cardiovascular Disease in Healthy General Populations of Young People (12–25 Years of Age)

A Scientific Statement From the American Heart Association and the American College of Cardiology

Endorsed by the Pediatric and Congenital Electrophysiology Society and American College of Sports Medicine

The theoretical aspiration to screen the entire 12- to 25-year-old population of the United States f[redacted]diovascular disease with ECGs would be an un[redacted]king of enormous magnitude, with massive resource demands, in

[redacted]

(16,27,39,43,62,77,110). In addition, the question has arisen of whether such a mass screening program with ECGs is ethically defensible if confined to only 1 segment of the population when others may also be at risk.

[redacted]

tion. The infrequency of these events in no way mitigates their importance or impact on families and the community. However, it should also be underscored that the unexpected nature of these tragedies on the athletic field magnifies the public perception of their incidence, particularly when a highly visible athlete is involved.

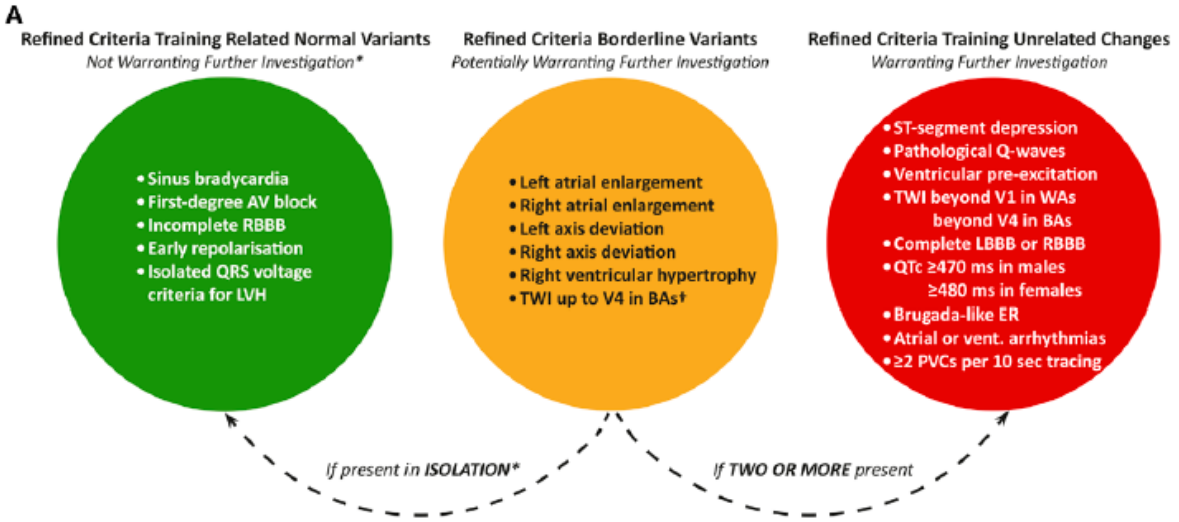
Table 1. ECG Parameters Used to Define Various ECG Abnormalities in the European Society of Cardiology Recommendations, Seattle Criteria, and Refined Criteria

ECG Abnormality	European Society of Cardiology Recommendations[9]	Seattle Criteria[12]	Refined Criteria
Left atrial enlargement	Negative portion of the P wave in lead V_1 ≥0.1 mV in depth and ≥40 ms in duration	Prolonged P wave duration of >120 ms in lead I or II with negative portion of the P wave ≥1 mm in depth and ≥40 ms in duration in lead V_1	As ESC
Right atrial enlargement	P-wave amplitude ≥2.5 mm in lead II, III, or aVF	As ESC	As ESC
Left QRS axis deviation	−30° to −90°	As ESC	As ESC
Right QRS axis deviation	>115°	>120°	As ESC
Right ventricular hypertrophy	Sum of R wave in V_1 and S wave in V_5 or V_6 ≥10.5 mm	Sum of R wave in V_1 and S wave in V_5 >10.5 mm and right axis deviation >120°	As ESC
Complete LBBB	QRS ≥120 ms, predominantly negative QRS complex in lead V_1 (QS or rS), and upright monophasic R wave in leads I and V_6	As ESC	As ESC
Complete RBBB	RSR′ pattern in anterior precordial leads with QRS duration ≥120 ms	Not relevant	As ESC
Intraventricular conduction delay	Any QRS duration >120 ms including RBBB and LBBB	Any QRS duration ≥140 ms or complete LBBB	As ESC
Pathological Q-wave	>4 mm deep in any lead except III, aVR	>3 mm deep or >40 ms duration in ≥2 leads except III and aVR	≥40 ms in duration or ≥25% of the height of the ensuing R wave
Significant T-wave inversion	≥2 mm in ≥2 adjacent leads (deep) or "minor" in ≥2 leads	>1 mm in depth in ≥2 leads V_2–V_6, II and aVF, or I and aVL (excludes III, aVR, and V_1)	As Seattle
ST-segment depression	≥0.5 mm deep in ≥2 leads	As ESC	As ESC
Ventricular preexcitation	PR interval <120 ms with or without delta wave	PR interval <120 ms with delta wave	As Seattle criteria

LBBB indicates left bundle-branch block; and RBBB, right bundle-branch block.

Sheikh, Circ, 2014

Sheikh, Circ, 2014



B

ESC Group 1 Training Related Changes
• Sinus bradycardia
• First-degree AV block
• Incomplete RBBB
• Early repolarisation
• Isolated QRS voltage criteria for LVH

ESC Group 2 Training Unrelated Changes	
• T-wave inversion	• Ventricular pre-excitation
• ST-segment depression	• Complete LBBB or RBBB
• Pathological Q-waves	• Long QT >440 ms in males
• Left or right atrial enlargement	• Long QT>460 ms in females
• Left axis deviation / left anterior hemiblock	• Short QT interval <380 ms
• Right axis deviation / left posterior hemiblock	• Brugada-like ER
• Right ventricular hypertrophy	• Atrial/ventricular arrhythmias

C

Seattle Criteria Abnormal Findings in Athletes		
• T-wave inversion beyond V2 in WAs and V4 in BAs†	• Left atrial enlargement	• Short-QT interval ≤320 ms
• ST-segment depression	• Right atrial enlargement	• Brugada-like ECG pattern
• Pathological Q-waves	• Right ventricular hypertrophy pattern	• Profound sinus bradycardia (<30 bpm)
• Complete left bundle branch block	• Ventricular pre-excitation	• Atrial tachyarrhythmias
• Intraventricular conduction delay *(any QRS ≥140 ms)*	• Long-QT interval ≥470 ms in males	• Premature ventricular contractions
• Left axis deviation	• Long-QT interval ≥480 ms in females	• Ventricular arrhythmias

KEY

AV: Atrioventricular
BAs: Black athletes
ER: Early repolarisation
ESC: European Society of Cardiology
LBBB: Left bundle branch block
LVH: Left ventricular hypertrophy
PVCs: Premature ventricular complexes
RBBB: Right bundle branch block
Sec: Second
TWI: T-wave Inversion
Vent.: Ventricular
WAs: White athletes

*In otherwise asymptomatic athletes with no family history or abnormal examination findings. †When preceded by characteristic convex ST-segment elevation.

Figure 1. The definition of an abnormal ECG using the (**A**) refined criteria, (**B**) European Society of Cardiology (ESC) recommendations,[9] and (**C**) Seattle criteria.[12]

HOW DO WE DEFINE COST EFFECTIVE?

- Tough question when the bottom line is possibly preventing sudden death in a young patient
  - How much dollar worth do you place on a single life
  - It comes down to the gamble of the risk of someone dying versus an acceptable expenditure

DOING THE MATH

Assumes 10 million Middle and High School athletes

CMS reimbursements:

Personal & family hx with physical = $25

ECG = $50

Subtotal = $750 Million

Positive Hx or PE in 15% results in 2nd Hx and PE ($100) with echo ($400)

Subtotal = $750 Million

Total = $1.5 Billion (yes, with a "B") per year

Total doesn't include administrative and operating costs, plus other additional medical expenses to add another $500 million

This comes to $330,000 per athlete with suspected relevant cardiac disease

The cost of preventing each theoretic death is $3.4 million

Maron, Circ 2007

Where should we devote most of our resources?

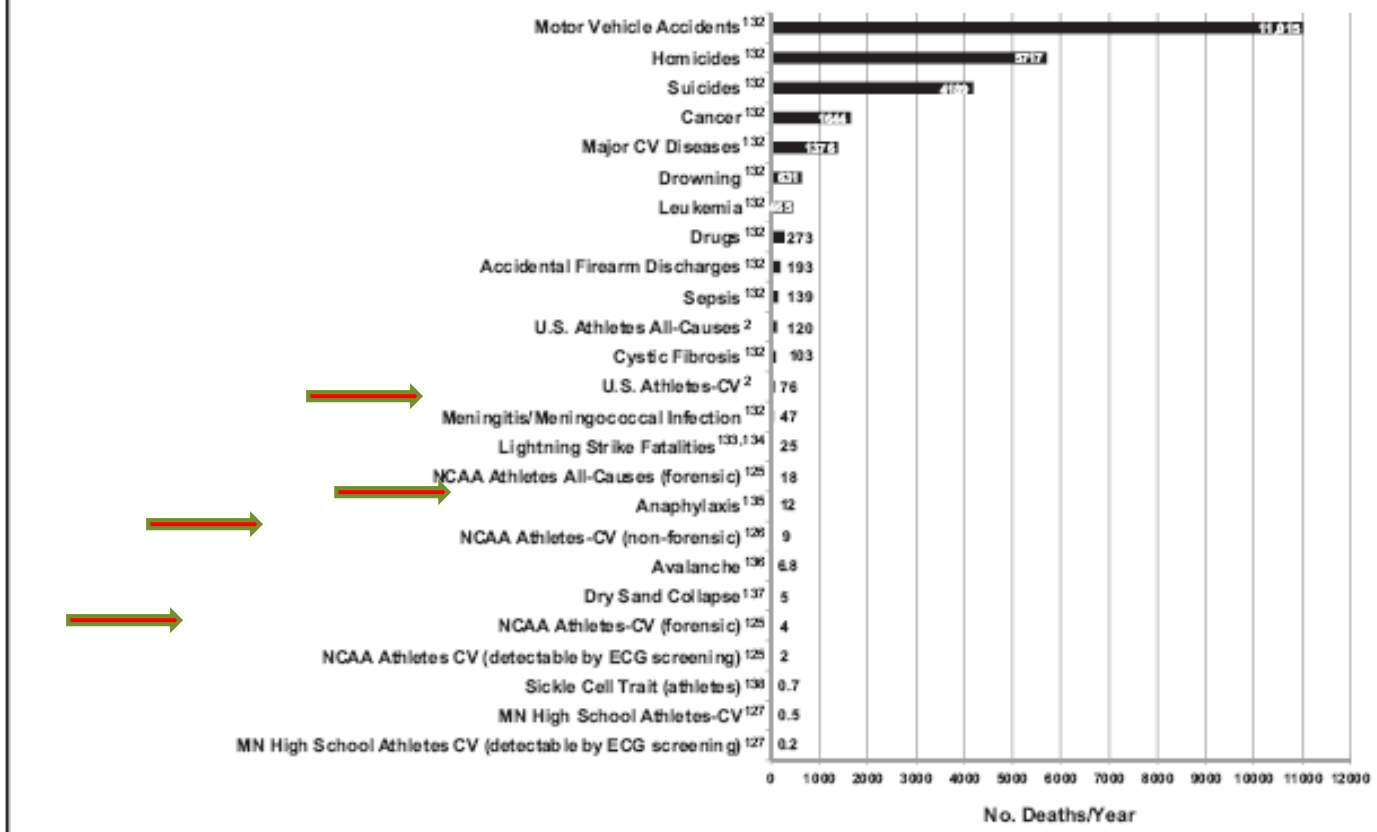


FIGURE Comparative Frequencies of Death Attributable to All Causes in Young Individuals Aged <25 Years

CV indicates cardiovascular; MN, Minnesota; and NCAA, National Collegiate Athletic Association.

From Maron, JACC, 2014

SUMMARY

- There are critical measures to take following sudden cardiac arrest
  - History, physical, family hx is crucial
  - Blood sample for genetic testing
- Congenital heart disease, channelopathies, and cardiomyopathies elevate the risk of sudden death
- Controversy remains regarding the use of ECGs to assess athletes